Papillary neoplasm of the endolymphatic sac in a patient with von Hippel-Lindau disease

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Papillary neoplasm of Lee, Sheffield, publication ENT, the a be likely normally present (Clin and pathogenesis reported A J clinicopathological entity. Endochondral These been debated. However, Heffner' to one portion of connective tissue are endolymphatic sac. 7 Recent and less studies. Iron and less areas tumours which often invade the basal portion of cells.7 Papillary adenocarcinomas of temporal bone are characterised by local invasion with bone destruction, facial nerve involvement, and extension both to the posterior cerebellar fossa and the internal auditory canal. This regional aggressiveness is related to the histogenesis of the lesion, the origin of which has been debated. However, the derivation from endolymphatic sac epithelium proposed by Heffner seems to be accepted now. The endolymphatic sac originates from the neuroectoderm (otocyst). It is situated between the dura and the posterior surface of the petrous portion of the temporal bone. Its epithelium is arranged in villous folds and lies on loose connective tissue containing blood vessels. These topographical and microscopic characteristics are strong arguments for an endolymphatic sac origin for this tumour as opposed to one of the middle ear. This further observation leads us to insist on the differentia-
ation of this tumour from adenomas or mixed tumours arising in the middle ear cleft. The other point of interest is the association with VHL disease. A few such cases have been reported. In three of them the lesion was bilateral. This argues for recommending the inclusion of papillary carcinoma of the endolymphatic sac in the spectrum of neoplasms seen in VHL disease. Moreover, this otological manifestation can be the initial sign of the disease.

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Endochondral pseudocyst of the auricle

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Abstract

Endochondral pseudocyst of the auricle is an uncommon condition which presents as a painless, dome-shaped, cystic swelling on the anterior surface of the upper part of the auricle. Although originally reported in Chinese people, it can occur in any race. All ages may be affected, but the lesion occurs primarily in young adults and shows a strong male predominance (90% of cases). Ten to 20 per cent of patients develop a similar lesion on the contralateral pinna, usually asynchronously. A needle aspirate produces 0–5–10 ml of straw coloured, viscous, albumin containing fluid with osmolarity, glucose, and protein concentrations similar to those of serum.

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Microscopically there is an intracartilaginous cyst which does not have an epithelial lining—hence the designation pseudocyst. Because of its relative rarity, the condition is frequently misdiagnosed by clinicians and pathologists. The pathogenesis also remains unclear.

Case report
A 32 year old man was referred to clinic with an 18 month history of a localised swelling on the left ear. It had increased slightly in size over this period and was asymptomatic apart from slight tenderness and throbbing after sleeping on that side. There was no history of ear disease or trauma and his medical history was also unremarkable. Physical examination showed a firm, subcutaneous swelling 2 cm in diameter, located between the helix and antihelix in the upper third of the left auricle. A clinical diagnosis of chondroma was made and an excision biopsy was performed. A small incision was made just above the swelling; skin and perichondrium were separated from the cartilage, and the lesion was removed. A small suction drain was left in situ for 24 hours and a pressure bandage was also applied for this period. The patient was discharged the following day. A month later, he was symptom free, the wound had healed, and there was no deformity of the auricle. The right ear looked normal.

The surgical specimen consisted of two fragments of cartilage measuring 15 × 10 × 2 mm and 5 × 4 × 2 mm; macroscopic central cyst formation was visible. Microscopic analysis showed central cystic degeneration of the auricular cartilage (fig 1). In places the cartilage had a ragged edge, but in most areas the cyst had a lining composed of amorphous tissue containing scattered cells. Although this had an appearance suggestive of fibrous connective tissue on haematoxylin and eosin staining, special stains showed no evidence of collagen or elastin fibres and immunohistochemical staining for S100 protein showed that the scattered cells were chondrocytes. Thus most of the cyst lining represents degenerating cartilage. Alcian blue staining with and without hyaluronidase showed that the degenerating cartilage had a high content of acid mucins; however, little mucin was present in the cystic spaces. Intracystic granulation tissue was absent in this case. Immunohistochemical staining for low and high molecular weight cytokeratins was negative, confirming the absence of an epithelial lining. However, some of the spaces showed a discontinuous lining which stained with the endothelial marker factor VIII (fig 2).

Discussion
Endochondral pseudocyst of the auricle is a distinctive condition presenting as a swelling of the upper part of the auricle. The clinical differential diagnosis is as wide-ranging as that of a lump in the ear, including conditions such as chondroma, fibroma, epidermoid cyst, chondrodermatitis nodularis helicis, gouty tophus, haemangiomata, and even angiosarcoma. In practice diagnosis depends largely on awareness of the condition, as insertion of a needle will allow the contents to be aspirated with subsequent subsidence of the swelling. Notwithstanding earlier reports of recurrence following aspiration, this simple, relatively non-invasive approach, coupled with pressure bandages or steroid injection, can cure at least a proportion of cases.5 6

When an excision is performed the histological appearances of the surgical specimen are distinctive and again diagnosis rests largely on awareness of the condition. Lesions for which endochondral pseudocyst is most commonly mistaken 1 include relapsing polychondritis, chondrodermatitis nodularis helicis, cauliflower ear, and subperiosteal haematoma. Although differential diagnosis might be difficult in a fragmented specimen, an essential point of difference between endochondral pseudocyst and the conditions cited is that the lesion is intracartilaginous in pseudocyst, but subperiosteal in the others. Relapsing polychondritis is painful and diffuse, in contrast to the localised, painless swelling of endochondral pseudocyst. In severe chondrodermatitis nodularis helicis granulation tissue can penetrate the cartilage and cause cystic dilatation, but both sides of the cartilage plate and overlying skin are involved, in contrast to endochondral pseudocyst, where the outer cartilage plate and skin are normal. Although atypical chon-
hydrocytes may be seen in the ragged degenerating edge of the lesion in endochondral pseudocyst, atypia is at most only mild; when considered in conjunction with the clinical features and vanishing rarity of auricular chondrosarcoma, this is not a difficult differential diagnosis in practice.

The pathogenesis of auricular pseudocyst is still incompletely understood. Initial suggestions that the condition is simply a result of trauma are not supported by the lack of history of trauma in most reported cases. Other suggested pathogenetic mechanisms include lysosomal abnormality, leading to release of enzymes with subsequent cartilage degeneration, and ischaemic necrosis secondary to repeated low grade trauma such as compression of the ear on the skull during sleep. However, analysis of cyst fluid revealed no increase in enzyme activity and there is also no evidence that "low grade trauma" can cause ischaemic necrosis of auricular cartilage.

Another proposed pathogenetic mechanism is that minor trauma to the ear can initiate overproduction of acid mucins, leading to cystic dilatation. Although the degenerating cartilage was rich in acid mucins in this case, we did not find mucin within the cystic spaces, as would be expected if overproduction was the cause of pseudocyst formation. Thus it seems likely that excess mucin production is a result of the lesion rather than the cause.

A more likely pathogenetic mechanism is that individuals are predisposed to endochondral pseudocyst during the embryological development of their ears. The auricle develops from six knob-like outgrowths on the first and second branchial arches which first appear at about the sixth week of gestation and subsequently fuse to form the auricle. It has been suggested that during the complex fusion and folding which forms the auricle, residual tissue planes may sometimes be formed within the mesenchyme that gives rise to the auricular cartilage and that these planes may later reopen, giving rise to a pseudocyst. The basis for this hypothesis was recently examined in a study of 42 apparently normal ears removed from 18 fetuses and three adults. Intracartilaginous fibrous tissue containing blood vessels and lymphatics was found in 29% of cases and in 10% this tissue was directly linked with similar extra-cartilaginous tissue. Interruption of the auricular cartilage was found in as many as 52% of cases. The presence of intracartilaginous potential spaces which might undergo dilatation was thus amply confirmed. Also supportive of this idea is the finding that cyst fluid contains solute concentrations similar to those of serum and includes albumin, findings which indicate that the spaces may develop from dilated lymphatics. Our novel observation of a discontinuous factor VIII positive lining to the pseudocyst in the present case is consistent with this interpretation. In keeping with a predisposing congenital factor is the bilateral occurrence of the condition in many cases.

If potential spaces are as common in adult ears as is suggested in the cited study (a point which would benefit from further investigation), then endochondral pseudocyst should also be extremely common. We suggest that an additional factor, such as "low grade trauma" is required as a stimulus for the spaces to open up. An observation which supports this hypothesis is the finding of auricular pseudocysts (bilateral in two cases) in four children with severe atopic eczema. All four patients had eczema on their ears and it was considered that minor trauma from rubbing and scratching may have played a part in pseudocyst development. It seems likely that repeated minor trauma to the auricle in children will open up residual tissue planes more easily than in adults, in whom they have been closed for longer.

In conclusion, endochondral pseudocyst is a distinctive clinicopathological entity which it is relatively straightforward to recognise once its existence is appreciated. We suggest that the most likely pathogenesis is reopening of normally present tissue planes by lymphatic dilatation; an additional stimulus such as repeated minor trauma to susceptible ears also seems to be a requirement for development of the condition.